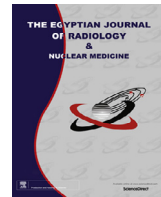




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Case Report

Primary hyperparathyroidism in a child: The musculoskeletal manifestations of a late presenting rare endocrinopathy



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ABSTRACT

Primary hyperparathyroidism (PHPT) is rare in children and adolescents, but has greater morbidity in this age group. Most of these patients show predominantly skeletal pathology and to a lesser extent renal involvement. Osteopenia, osteoporosis and subperiosteal resorption are frequently encountered radiographic skeletal signs. This study describes the orthopedic manifestations of PHPT in a child. PHPT in this child exhibited a late presentation with significant clinical morbidity and extensive radiographic manifestations. The characteristic radiographic pattern of PHPT in childhood is an important contributor to the diagnosis. The radioclinical and biochemical correlations augment diagnostic accuracy and delineate extent of skeletal pathology.

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1. Introduction

Primary hyperparathyroidism (PHPT) in children is a rare entity. In contrast with the clinical profile of PHPT in adults, PHPT is much less common in infants and children, with an incidence estimated at only 2–5 in 100,000 [1]. Primary hyperparathyroidism appears to be a more aggressive disorder in children than in adults. In most cases, PHPT will result from a single benign parathyroid adenoma [1–3]. Childhood and adolescent PHPT often presents with vague symptoms. These symptoms usually include bone

pain and abdominal pain. Most of these patients show predominantly skeletal pathology and to a lesser extent renal involvement. Interestingly, PHPT is clinically symptomatic in most younger patients. Few, if any, young patients with PHPT are discovered incidentally to have asymptomatic hypercalcemia [2–5]. Osteopenia, osteoporosis and subperiosteal resorption are the most often encountered radiographic skeletal signs. Primary hyperparathyroidism affects compact bone more than trabecular bone with particular sensitivity in the cortices of long bones [1–5]. We assume that describing consistent radiographic features of PHPT may aid diagnostic accuracy and help delineate extent of skeletal involvement. The purpose of this study was to describe the orthopedic manifestations of PHPT in a child. The clinical orthopedic profile was correlated to the skeletal radiographic characteristics of the patient with literature update.

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2. Case report

2.1. Clinical presentation

A 13 year old girl presented to our pediatric orthopedic outpatient clinic. The presenting symptom was generalized bone aches, especially of the spine and lower extremities. Repeated fractures arising from trivial trauma were reported. The patient's symptoms deteriorated over the past 6 years. Prior to presentation to the authors the patient received repeated simple cast immobilization and analgesics. She was presumably treated as a case with traumatic fractures or osteogenesis imperfecta. Eventually the patient became wheelchair ridden for the past year. No history of underlying disease or surgical interventions was reported. No family history suggestive of multiple endocrine neoplasia was encountered. General examination revealed normal parameters. The patient exhibited an average built and had already achieved menarche at time of presentation. Local examination revealed diffuse musculoskeletal tenderness, especially of the lower limbs and

spine. There was painful restriction of the range of motion of the affected joints. An Informed written consent was obtained from the patient and her parents for being included in the study. The authors declare that no conflict of interest exists. No financing was received for this study.

2.2. Imaging findings

The patient received orthogonal plain radiographic examination over the pelvis, whole spine and legs to characterize and evaluate the extent of the disease. The images depicted a wide array of manifestations attributed to generalized skeletal demineralization, bone formation and pathologic fractures (Figs. 1–3).

2.3. Laboratory findings

Laboratory findings revealed hypercalcemia 11.8 mg/dl (range 9.5–10.4)/2.92 mmol/L (range 2.38–2.60), elevation of the alkaline phosphatase 786 IU/L (range 153–362)/13.13 μ kat/L (range 2.56–6.05), serum phosphorus

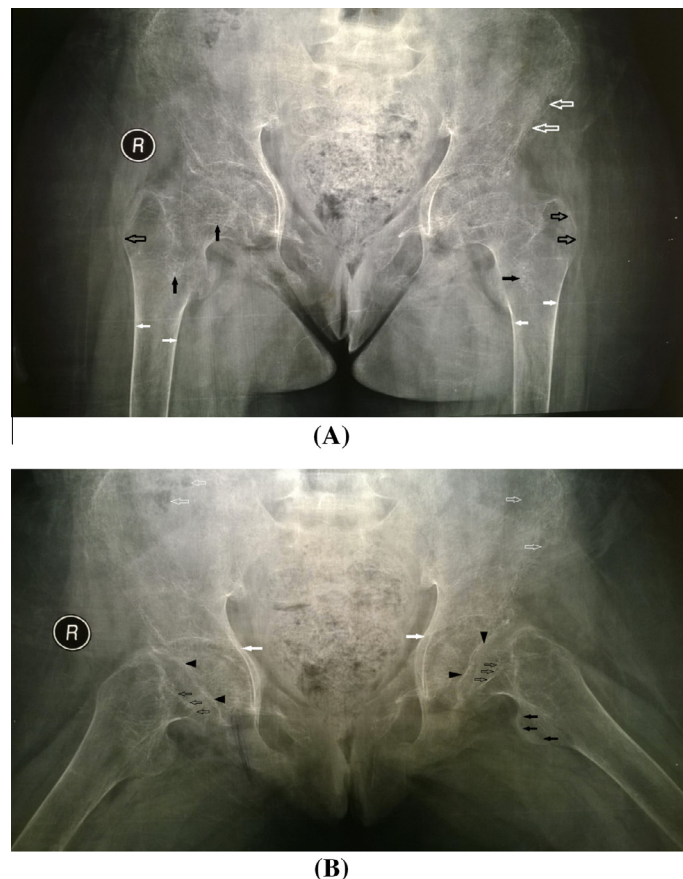


Fig. 1. (A and B): Plain radiographs of the pelvis and both hips anteroposterior view (A): Note the diffuse demineralization and trabecular resorption, coarse appearance of the trabeculae (solid black arrows), subtendinous resorption of greater trochanters (hollow black arrows), subperiosteal resorption of the iliac bone (hollow white arrows), and cortical thinning (white solid arrows). Plain radiographs of the pelvis and both hips lateral view (B): Note the diffuse skeletal rarefaction, the thin sclerotic lines surrounding a very faint lucent line over the femoral necks mostly representing a pathologic stress fracture (hollow black arrows), premature fusion of the proximal femoral physis (black arrow heads), subtendinous resorption of the lesser trochanter (black arrows), deepening of the acetabular concavity and thinning of its medial wall (solid white arrows), subperiosteal and intramedullary resorption of the iliac bone (hollow white arrows).

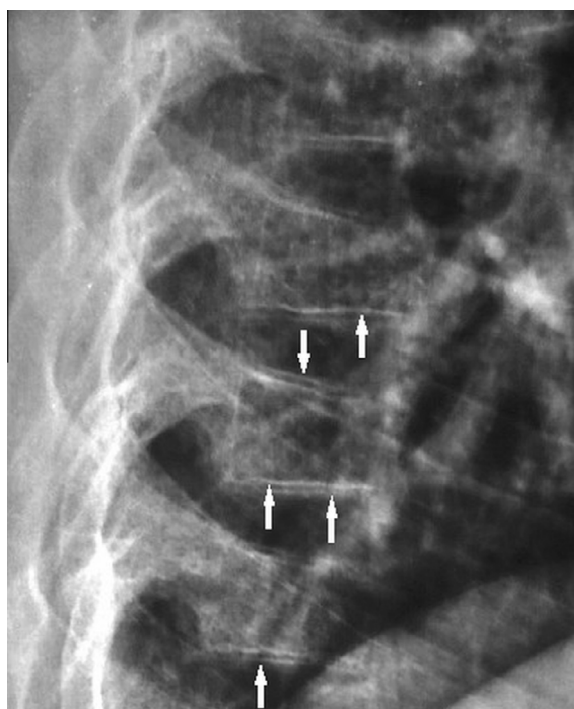


Fig. 2. Plain radiographs of the dorsal spine lateral view. Note the generalized osteopenia, end plate sclerosis and adjacent subchondral sclerosis (white arrows), and anterior vertebral body wedging.

2.8 mg/dl (range 3.5–4.9)/0.90 mmol/L (range 1.13–1.58), and an elevated serum parathyroid hormone 891 ng/L (range 15–65). Serum creatinine and urine analysis revealed normal findings. The diagnosis of primary hyperparathyroidism was established by correlating the clinical, radiologic and laboratory findings of the patient.

3. Discussion

Plain radiographs may yield the most specific findings that are consistent with the PHPT, and radiography is the preferred examination when the clinical findings suggest primary hyperparathyroidism. Furthermore radiography may be useful in defining the extent of damage [3,5]. Hyperparathyroidism is a disease of increased bone resorption and bone formation. Subsequently, plain radiographic findings may include resorption and sclerosis of numerous sites in the skeletal system. Primary hyperparathyroidism affects compact bone more than trabecular bone with particular sensitivity in the cortices of long bones leading to subperiosteal bone resorption (seen as periosteal elevation on plain radiography). In advanced PHPT, the entire skeleton can be involved [1,4,5]. The pattern of skeletal demineralization depicted on radiographs of the current case conforms to observations of the previous authors. Primary hyperparathyroidism in childhood and adolescence is usually diagnosed later, and presents with clinically significant morbidity [1–4]. Advanced bone changes tend to be common. It has been emphasized that the presenting symptoms of PHPT in childhood and adolescents are



Fig. 3. Plain radiographs of both legs anteroposterior view. Note the complete cortical disruption; pathologic fracture of the proximal medial cortex of the left leg (hollow black arrow), marked cortical thinning of both tibial shafts (white arrows), premature fusion of the proximal and distal tibial growth plates (solid black arrows).

predominantly musculoskeletal [2–4]. Osteopenia, osteoporosis and subperiosteal resorption are the most often encountered radiographic skeletal signs described [1–5]. The clinical profile of the current case is in line with that of the previous authors, especially the delayed presentation and the predominately skeletal complications. The current study demonstrates a clear correlation between the extensive bone resorption, cortical thinning and pathologic fractures found on the plain radiographs and the patient's severe bone pain, tenderness and inability to weight bear.

The longstanding patient immobilization may in part be a contributor to the generalized bone rarefaction seen on plain radiographs. Premature fusion of the major growth plates of the lower limbs and the other sclerotic manifestations may be attributed to the hypercalcemia and bone formation phase, especially in the early stage of the patient's longstanding disease course. Brown tumors of the long bones and a salt-and-pepper appearance of the skull occur in less than 5% of the United States patients with primary disease [6]. Brown tumors were not reported in the presented case.

Another way to monitor the severity of bony involvement is with bone densitometry, determined by dual energy X-ray absorptiometry (DEXA). Bone densitometry is the preferred diagnostic modality for the evaluation of osteoporosis, which is one of the most common findings

in patients with PHPT. Bone density in the hip and lumbar spine, for which pediatric reference range values are often integrated into the computer software of the machine, is expected to be low compared with age-related reference range values [6]. However, osteoporosis may be associated with other diagnoses; therefore, the specificity of PHPT may be limited. Because of the obvious radiographic skeletal manifestations found in the presented case, both in extent and severity, it was assumed that added value from DEXA may be minimal. As described, children and adolescents with PHPT are usually diagnosed later and a few if any young patients with PHPT are discovered incidentally to have asymptomatic hypercalcemia. We suggest that DEXA may be a diagnostic indicator in these incidentally discovered patients prior to the establishment of manifest radiographic findings.

3.1. Conclusion

Primary hyperparathyroidism in children and adolescents seems to be a disease with significant skeletal morbidity. The current pediatric case study showed a clear correlation between the clinical and radiographic skeletal manifestations of PHPT. Skeletal radiographs were helpful to visualize the extent and multiplicity of the lesions associated with PHPT. In addition to diffuse osteopenia and skeletal lesions presented in this case lesions were specifically bilateral, symmetric and multifocal, exhibiting different types of bone resorption. Furthermore, the coexistence of resorption and sclerosis was found at numerous sites in

the skeletal system. In addition to the clinical and biochemical profile, the characteristic pattern of involvement on plain radiographs augmented the diagnostic accuracy. Early diagnosis of PHPT in children and adolescents is fundamental to avoid disease related complications and initiate timely and appropriate treatment.

Conflict of interest

The authors of the current study declare that no conflict of interest exists. No financing was received for research on which our study is based.

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